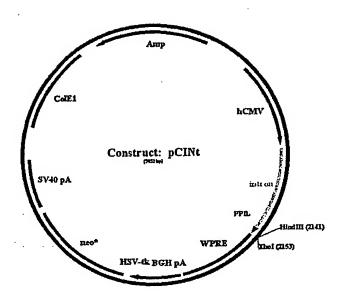
FIGURE 1

GAAF: 5'-GCGATAGGTACCGCCATGGGAGTGAGGCACCCGCCCTGCTCCC-3'

GAAR: 5'-GCGATACTCGAGTCAACACCAGCTGACGAGAAACTGCTCTCCC-3'

FIGURE 2



100

FIGURE 3

atgaaagggtccctcctgctgctgctggtgtcaaacctgctcctgtgccagagcgggtcc M K G S L L L L V S N L L L C Q S G S qqaqccggggcccacatcctactccatgatttcctgctggttccccgagagctgagtggc G A G A H I L L H D F L L V P R E S P V L E E T H P A H Q Q G A S ccccgggatgcccaggcacaccccggccgtcccagagcagtgcccacacagtgcgacgtc RDAQAHPGRPRAVPTQCDV cccccaacagccgcttcgattgcgcccctgacaaggccatcacccaggaacagtgcgag PPNSRFDCAPDK·AI TQEQCE gcccgcggctgctgctacatccctgcaaagcaggggctgcagggagcccagatggggcag A R G C C Y I P A K Q G L Q G A Q M G Q ccctqqtqcttcttcccacccaqctaccccaqctacaaqctqqaqaacctqaqctcctct PWCFFPPSYPSYKLENL gaaatgggctacacggccaccctgacccgtaccacccccaccttcttccccaaggacatc MGYTATLTRTTPTFFKD ctgaccctgcggctggacgtgatgatggagactgagaaccgcctccacttcacgatcaaa T L R L D V M M E T E N R L H gatccagctaacaggcgctacgaggtgcccttggagaccccgcgtgtccacagccgggca ANRRYEVPLETPRVH ccgtccccactctacagcgtggagttctccgaggagcccttcggggtgatcgtgcaccgg PLYSVEFSEEPFGVIVHR cagctggacggcgcgtgctgctgaacacgacggtggcgcccctgttctttgcggaccag Q L D G R V L L N T T V A P L F F A D Q. ttccttcagctgtccacctcgctgccctcgcagtatatcacaggcctcgccgagcacctc L Q L S T S L P S Q Y I T G L A E H agtcccctgatgctcagcaccagctggaccaggatcaccctgtggaaccgggaccttgcg PLMLSTSWTRITLWNRDLA cccacgcccggtgcgaacctctacgggtctcaccctttctacctggcgctggaggacggc P T P G A N L Y G S H P F Y L A L E D G gggtcggcacacggggtgttcctgctaaacagcaatgccatggatgtggtcctgcagccg G S A H G V F L L N S N A M D V V L Q P agccctgcccttagctggaggtcgacaggtgggatcctggatgtctacatcttcctgggc PALSWRSTGGILDVYIF ccagagcccaagagcgtggtgcagcagtacctggacgttgtgggatacccgttcatgccg E P K S V V Q Q Y L D V V G Y P F M P ccatactggggcctgggcttccacctgtgccgctggggctactcctccaccgctatcacc WGLGFHLCRWGYSST A I cgccaggtggtggagaacatgaccagggcccacttccccctggacgtccaatggaacgac D V Q W N RQVVENMTRAHFPL ctggactacatggactcccggagggacttcacgttcaacaaggatggcttccgggacttc D Y M D S R R D F T F N K D G F R D F ccggccatggtgcaggagctgcaccagggcggccggcgctacatgatgatcgtggatcct A M V Q E L H Q G G R R Y M M I gccatcagcagctcgggccctgccgggagctacaggccctacgacgagggtctgcggagg S S S G P A G S Y R P Y D E G L R ggggttttcatcaccaacgagaccggccagccgctgattgggaaggtatggcccgggtcc ETGQPLIGKVW V F I \mathbf{T} N actgccttccccgacttcaccaaccccacagccctggcctggtgggaggacatggtggct

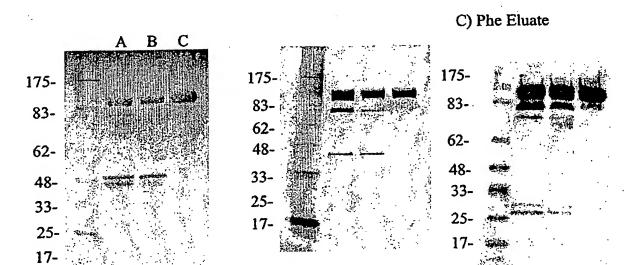
D M V A PTALAWWE F \mathbf{T} N D \mathbf{T} A F Р gagttccatgaccaggtgcccttcgacggcttgtggattgacatgaacgagccttccaac F H D Q V P F D G L W I D M N E P I R G S E D G C P N N E L E N P cctggggtggttggggggaccctccaggcggccaccatctgtgcctccagccaccagttt G V V G G T L Q A A T I C A S SHQF ctctccacacactacaacctgcacaacctctacggcctgaccgaacccatcgcctcccac S T H Y N L H N L Y G L T E P I A S H agggcgctggtgaaggctcgggggacacgcccatttgtgatctcccgctcgacctttgct RALVKARGTRPFVISRSTF ggccacggccgatacgccggccactggacgggggacgtgtggagctcctgggagcagctc G H G R Y A G H W T G D V W S S W E Q L gcctcctccgtgccagaaatcctgcagtttaacctgctgggggtgcctctggtcggggcc S S V P E I L Q F N L L G V P L V DVCGFLGNTSEELCVRW ggggccttctaccccttcatgcggaaccacaacagcctgctcagtctgccccaggagccg G A F Y P F M R N H N S L L S L P Q E tacagcttcagcgagccggcccagcaggccatgaggaaggccctcaccctgcgctacgca Y S F S E P A Q Q A M R K A L T L R Y A ctcctcccccacctctacacactgttccaccaggcccacgtcgcgggggagaccgtggcc P H L Y T L F H Q A H V A G E cggcccctcttcctggagttccccaaggactctagcacctggactgtggaccaccagctc RPLFLEFPKDSSTWTVD H Q L ctgtgggggggggccctgctcatcaccccagtgctccaggccgggaaggccgaagtgact LWGEALLITPVLQAGKAEVT ggctacttccccttgggcacatggtacgacctgcagacggtgccaatagaggcccttggc G Y F P L G T W Y D L Q T V P I E A L G agcctcccaccccacctgcagctccccgtgagccagccatccacagcgaggggcagtgg L P P P P A A P R E P A I H S E G Q W gtgacgctgccggccccctggacaccatcaacgtccacctccgggctgggtacatcatc TLPAPLDTINVHLRAGY ccctgcagggccctggcctcacaaccacagagtcccgccagcagcccatggccctggct L Q G P G L T T T E S R Q Q P M A gtggccctaaccaagggtggagaggcccgaggggagctgttctgggacgatggagagagc T K G G E A R G E L F W D D G ctggaagtgctggagcgagggcctacacacaggtcatcttcctggccaggaataacacg L E V L E R G A Y T Q V I F L A R N atcgtgaatgagctggtacgtgtgaccagtgagggagctggcctgcagctgcagaaggtg SEGAGLQLQK IVNELVRVT actgtcctgggcgtggccacggcgccccagcaggtcctctccaacggtgtccctgtctcc G V A T A P Q Q V L S N G V P V aacttcacctacagccccgacaccaaggtcctggacatctgtgtctcgctgttgatggga F T Y S P D T K V L D I C V S L L M G gagcagtttctcgtcagctggtgttga E Q F LVSW C

A) Blue Eluate

B) Q Eluate

5/8

FIGURE 4



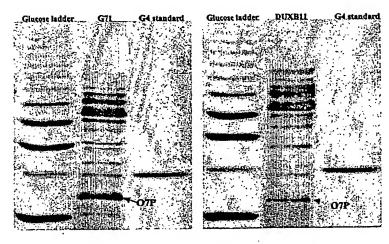
Ag+ 5ug BM103/lane

Commassie 5ug BM103/lane

Western (1:5000) Primary 0.5ug BM103 / lane

FIGURE 5

FACE profiles for GAA from G715 and DUXB11



O7P 19% of total oligosaccharide profile

O7P 6.7% of total oligosaccharide profile

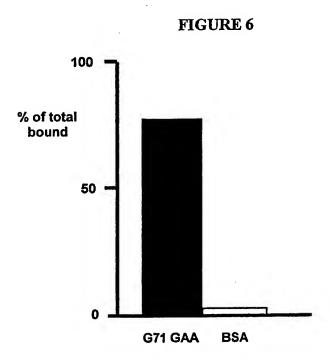


FIGURE 7

Kuptake for DUX rhGAA = 2.95 nM, Kuptake for G71 rhGAA = 1.31 nM

